6,7], p<0,0001). These associations remained significant after multiple logistic regression analysis including the severity of the heart defect (univentricular or biventricular physiology).

Conclusion: Women are more likely to terminate pregnancy if extracardiac or chromosomal anomalies are associated. Post natal survival is strongly influenced by these associated anomalies.

0524

Experience with foetal supraventricular arrhythmias

Francis Bessiere (1), Hervé Joly (1), Jérôme Massardier (2), Magali Veyrier (1), Nicolas Pangaud (1), André Bozio (1), Sylvie Di Filippo (1)
(1) Hôpital Cardiovasculaire, Cardiologie pédiatrique et congénitale, Lyon, France – (2) Hôpital Femme Mère Enfants, Obstétrique, Lyon, France

This study was to review experience and outcomes of supraventricular (SV) arrhythmias in fetus Methods: Cases were divided in groups: SVPB= premature SV beats, NSSVT= non-sustained SV tachycardia, SSVT= sustained SV tachycardia, and AF= atrial flutter. Heart failure (HF) was defined as foetal hydrops or isolated effusion (pericardial or pleural or ascitis). Outcome was favourable if arrhythmia resolved or stabilized until full-term birth, not-favourable if premature birth or foetal death occurred.

Results: 188 fetuses were included:89 in SVPB(47.3%), 31 in NSSVT(16.5%), 60 in SSVT(31.9%), 8 in AF(4.3%), aged at diagnosis 30.8±4.5weeks (no difference between groups). Foetus HR at diagnosis was 241±30bpm in SSVT vs 226±26 in AF. Antiarrhytmic therapy was administered in sustained tachycardia (83% of SSVT and 71% AF): 28 had 1 medication, 25: 2 medications, 2: 3 medications. Complication occurred in 29 cases, all in SSVT and AF (29 of 68= 43%): 18 hydrops, 5 ascitis, 4 pericarditis, 1 pleural effusion and 1 LVdysfunction+MR, was more frequent in SSVT (86%: hydrops in 30%) than AF (51%: no hydrops), p= 0.08. Fetal HF was associated with HR at diagnosis: 251±25bpm in hydrops vs 228±31bpm in no-hydrops (p=0.025). Outcome was favourable in SVPB and NSSVT, in 45 of SSVT+AF (79%). Tachycardia resolved in 36, more frequently in SSVT (57%) than AF (25%). HR only decreased in 9cases. Premature birth occurred in 10, foetal death in 2. Outcome was not associated with HR or weeks of gestation at diagnosis. Defavourable outcome was more frequent in hydrops or isolated effusion (57%) than in uncomplicated cases (10%, p= 0.0002). Resolution occurred in 45% hydrops vs 66% of non-hydrops cases. Digoxine decreased from 79% of cases before 2000 to 33% after 2000, while flecaine increased from 14% to 48.5%. There was no relationship between therapy or number of medications and outcomes.

Conclusion: Fetal SSVT more frequently resolves but has worse outcome than AF, especially if HR at diagnosis is high and hydrops occurs. Larger scale prospective studies are needed to evaluate the efficacy of flecaine compared to digoxine therapy.

0537

Risk markers of cardiac events in patients with Marfan syndrome diagnosed during childhood

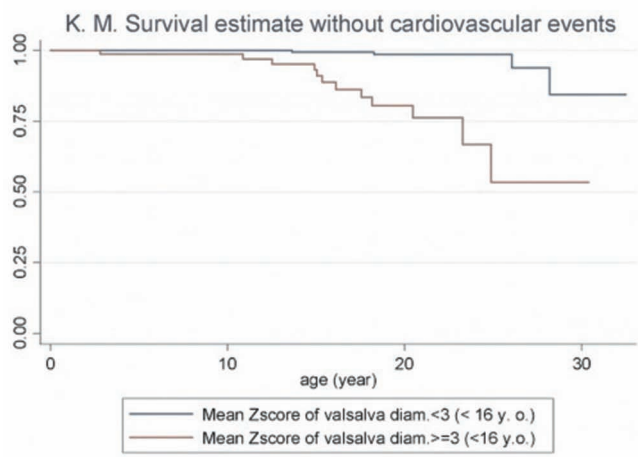
Sébastien Hascoët (1), Yves Dulac (2), Jean-Bernard Ruidavets (3), Thomas Edouard (4), Florence Arnoult (5), Olivier Milleron (5), Chantal Stheneur (6), Bertrand Chevallier (6), Cécile Zordan (7), Sylvie Odent (8), Nicole Philip (9), Laurence Olivier-Faivre (10), Bruno Leheup (11), Sophie Dupuis-Girod (12), Philippe Acar (2), Jean Ferrière (3), Guillaume Jondeau (5)
(1) CHU Toulouse, Hôpital des enfants, Pediatric cardiac surgery, Toulouse, France – (2) CHU Toulouse, Hôpital des enfants, Pediatric cardiology, Toulouse, France – (3) CHU Toulouse, Epidémiology, U 10 27, INSERM, Toulouse, France – (4) CHU Toulouse, Hôpital des enfants, Endocrinologie pédiatrique, Toulouse, France – (5) CHU Bichat-Claude Bernard-APHP, Cardiologie, Centre de référence pour le syndrome de Marfan et apparentés, Paris, France – (6) Hôpital Ambroise Paré-APHP, Pédiatrie, Boulogne, France – (7) CHU Bordeaux M3C, Pediatric and congenital cardiology, Bordeaux, France – (8) CHU Rennes, Hôpital Sud, Génétique, Rennes, France – (9) CHU la Timone enfant-APHM, Génétique clinique, Marseille, France – (10) CHU Dijon, Hôpital des enfants, Pédiatrie – Génétique clinique, Dijon, France – (11) CHU Nancy Brabois, Hôpital des enfants, Cardiologie pédiatrie, Vandoeuvre Les Nancy, France – (12) CHU Lyon, Hôpital Louis Pradel, Génétique clinique, Bron, France

Risk markers of cardio-vascular events in children with Marfan syndrome remain little known. Aortic root z-score measurements have been recently updated. We assessed the prognostic value of aortic root z-score in patients with Marfan syndrome diagnosed during childhood.

Methods: From the French multicenter database, 457 patients with Marfan syndrome, diagnosed before 18 y.o., without a history of cardiac event were prospectively included in this cohort study. Echocardiographic measurements of aortic root diameters were performed at each visit. We calculated the Z-score of aortic root measurements using the Bichat formula for each visit. Mean z-score was defined as the mean of the z-score (MeanZS15) calculated for each measurement before the age of 16.

Results: Median age at end of FU was 15.9 years (interquartile 10.9-20.3). FU was complete for 69.5% of patients. Median FU was 4.6 years. A cardiovascular event occurred in 17 patients (3.7%, prophylactic aortic surgery n=14, aortic dissection n=1 and deaths n=2). Survival free of cardiac events was 85.1% in patients with a meanZS15 of the Valsalva diameter <3 and 56.4% in patients with a meanZS15 of the Valsalva diameter ≥3 (p<0.0001 by log-rank test).In univariate analysis, Valsalva meanZS15 ≥3, age at inclusion in the database, a lower heart rate and an increased arm/height ratio were associated with an increasing risk of cardiac events (p<0.0001, p=0.04, p=0.01 and p=0.04 respectively). After multivariate adjustment using a cox proportional hazards model, only Valsalva meanZS15 ≥3 and lack of FBN1 or TGBB mutation identified were associated with an increasing risk of cardiac events (p<0.0001 and p=0.04 respectively).

Conclusion: Valsalva meanZS15 may help to predict whose children with a Marfan syndrome will have a cardio-vascular event. Therefore a particular attention to beta-blockade treatment observance and a regular follow-up may be suggested in high-risk children.



Abstract 0537 – Figure: k.m. survival estimate free from cardiac events

0564

Can systolic Doppler velocimetry of fetal aortic isthmus help predicting post-natal clinical impact of ventricular septal defects?

Julie Thomas-Chabaneix (1), Jean-Claude Fouron (2), Nagib Dahdah (2)
(1) CHU Bordeaux, Maladies cardiaques congénitales, Pessac, France – (2) CHU Ste Justine, Montréal, Canada

Objectives: Left ventricular ejection causes a forward flow in the fetal aortic isthmus while right ventricle (RV) has a retrograde influence. The objective of this study was to test the hypothesis that non restrictive ventricular septal defects (VSD) could cancel out the normal fetal RV preponderance and its retrograde systolic effect on the isthmic flow pattern.